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Nerve deafness in early syphilis

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That the nervous system is commonly invaded in secondary and sometimes in primary syphilis is well known and some abnormalities have been reported in 40–45 per cent. of spinal fluids (Stokes, Beerman, and Ingraham, 1944) although much lower figures (*e.g.* 5 per cent.) have been more recently described (Bauer, Price, and Cutler, 1952).

Sometimes frank early syphilitic meningitis may occur with headaches; optic neuritis may be encountered (Lorentzen, 1967) and the signs may resemble those of cerebral tumour (Huffmann, 1966). Although early invasion of the nervous system is usually asymptomatic, some investigators, using more detailed examination, have described a high incidence of neurological signs; these have included a widespread depression of pallaesthesia (Goldblatt 1953), and irregularity of the pupils, anisocoria, and diminished light reactions which have been noted by luminescent studies in a high proportion of patients (Braitsev and Kochetkov, 1968).

Nerve deafness is generally regarded as part of the congenital disease, but it may sometimes occur early in the acquired infection, as was well known in the days when syphilis was more common (Stokes and others, 1944). Since the disease has declined in prevalence this complication has tended to be lost sight of, and reports of the condition in past decades are scanty; one such case was recorded by Alergant (1965).

As syphilis is again increasing in importance in many areas, it is likely that more cases will arise. The present paper records three examples recently encountered in London.

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Basic information (Table I)

The three patients were all single, homosexual males

TABLE I *Basic information and mode of presentation*

Case no.	1	2	3
Date	March, 1966	December, 1967	October, 1968
Age (yrs) Marital status Country of origin	38 Single United Kingdom	31 Single United Kingdom	38 Single United Kingdom
Occupation and hobbies	Teacher Music lover	Self-employed Music student	Cinema projectionist Hi-fi addict
Previous history of venereal disease	None	NSU in 1964 and 1966 (WR, VDRL, and RPCFT negative)	None
Referred from	E.N.T. Dept.	S.2 Contact (already attended E.N.T. Dept.)	Eye Dept. (already attended E.N.T. Dept. elsewhere)
Symptoms	Deafness Tinnitus Loss of tone (5 wks)	Deafness Tinnitus Booming sounds Distortion (2 wks)	Aches in joints (8 mths) Blurred vision (2 mths) Woolly hearing, 'like through a blanket', no tinnitus (3 mths)
Previous history of ear trouble	None	Otitis when young	None

in their thirties, who were born in the United Kingdom. All were concerned with the appreciation of music either professionally or as a hobby. One of them had been treated for non-specific urethritis 1–3 years previously when the serological tests for syphilis were negative.

Presentation (Table I)
One patient was referred from the Ear, Nose, and Throat department, one was a contact of a male with secondary syphilis, and one was referred from the Ophthalmic department—having been found to be sero-positive on investigation of iritis. The two latter patients were also attending E.N.T. departments.

Two had symptoms of deafness and tinnitus of 2 and 5 weeks' duration, respectively. The first patient had noted loss of tone when attending symphony concerts, the music 'sounding like the Tijuana Brass' and the second experienced booming sounds and distortion. The third patient, who complained also of blurred vision for the previous 2 months on account of iritis, had experienced no tinnitus, but he described his hearing as woolly, 'like through a blanket' and it had been so for three months. This patient had also had aching joints for the past 8 months for which he had received indomethacin from his doctor.

Findings on examination (Table II)
All three had Rinne-positive perceptive deafness and in all the overt physical signs of secondary syphilis were slight although adenitis was a constant feature. Peri-anal lesions were noted in two, but *T. pallidum* was recovered from neither.

In all three cases the serological tests for syphilis were strongly positive and there were abnormalities in the cell count and protein content of the cere-

TABLE II *Findings on examination*

Case no.	1	2	3
Ears	Perceptive deafness	Perceptive deafness	Perceptive deafness
Signs of secondary syphilis	Peri-rectal ulceration (DG Neg.) Congested throat Adenitis + +	? Faint rash in oblique light Slight adenitis	Bilateral iritis ? Healed anal sore (DG neg.) ? Rash Adenitis + +
Serology	WR + + 1/128 VDRL + RPCFT +	WR + + 1/32 VDRL + RPCFT +	WR + + 1/40 VDRL + RPCFT +
Cerebrospinal fluid	Cells 15 WBC/c.mm. Protein 55 mg. (per cent.) Lange 0000000 WR + RPCFT +	Cells 43 WBC/c.mm. Protein 56 mg. (per cent.) Lange 11100000 WR – RPCFT –	Cells 90 WBC/c.mm. Protein 77 mg. (per cent.) Lange 3333210 WR + RPCFT +
Chest x-ray	Negative	Negative	Negative

TABLE III *Treatment*

Case no.	1	2	3
Possible contraindications	Nil	Penicillin sensitive (rash, 1957) Gilbert's disease	Nil
Corticosteroids	Prednisone 355 mg. over 12 days (start 60 mg.)	Prednisolone 277 mg. over 13 days (start 60 mg.)	Prednisolone 130 mg. over 11 days (start 40 mg.)
Penicillin build-up (500–500,000 units crystalline 4-hrly)	Yes	No	Yes
Subsequent penicillin	Pro Pen 1·2 m.u. daily for 5 days (total 6 m.u.) PAM 1·2 m.u. twice a week (total 12 m.u.)	None	Crystalline penicillin 500,000 u. four times a day for 10 days, plus probenecid G. 2g. daily
Other antibiotics	None	Cephaloridine 3·5 g. over 4 days Erythromycin 28 g. over 14 days Tetracycline 42 g. over 14 days	Cephaloridine 2 g. daily for 5 days
Side-effects	None	Hearing worse on 4th day: ? Herxheimer ? Cephaloridine ? Progression	Audiogram worse after 8 days Penicillin rash (10th day)

brospinal fluid. In two fluids there were also positive Wassermann reactions and RPCF tests, while the Lange curve was abnormal in one. In the nerve deafness of late congenital syphilis, the spinal fluid has previously been stated to be 'invariably normal' (Morton, 1955).

Treatment (Table III, previous page)

DRUGS USED

All three patients were treated with an 11 to 13 day course of corticosteroids commencing with a first daily dose of 40 to 60 mg. prednisone or prednisolone. This was given in an attempt to prevent a Herxheimer reaction rather than for curative reasons. The use of such treatment has been shown to produce some improvement in the deafness of late congenital syphilis (Hahn, 1964), although the improvement is often only one of discrimination and not of the pure tone levels (Patterson, 1968).

Two patients (Cases 1 and 3) were simultaneously given ten to eleven 4-hrly injections of crystalline penicillin commencing with 500 and increasing to 500,000 units. This was followed in Case 1 by five daily intramuscular injections of 1·2 m.u. procaine penicillin and then by ten twice weekly injections of 1·2 m.u. PAM. In Case 3, injections of 500,000 units crystalline penicillin were injected four times a day for 10 days and oral probenecid 500 mg. was also given four times a day. This regime was replaced by cephaloridine 1 g. intramuscularly twice daily for 5 days when a penicillin rash appeared on the 10th day.

The remaining patient (Case 2) had a history of earlier penicillin sensitivity and had also been suspected elsewhere to have Gilbert's syndrome (a congenital failure of bilirubin transport). He was therefore treated initially with cephaloridine, but after receiving 3·5 g. intramuscularly over 4 days his hearing, particularly in the right ear, became much worse, and the cephaloridine was discontinued. He was given instead 28 g. erythromycin orally over the next 14 days and later tetracycline 42 g. over the same period. It was uncertain whether the worsening was due to a Herxheimer effect in spite of the prednisolone, to the effects of cephaloridine, or to progression of the disease. A further lumbar puncture was performed and the cell count of the fluid had fallen to only 5/c.mm. so that progression seemed to be unlikely. The Dunlop Committee was notified but no similar cases involving cephaloridine had been reported to them. In retrospect, it is felt that it was probably a Herxheimer effect, for in Case 3, although no additional hearing impairment was noted clinically, the audiogram appeared to be less good 8 days from treatment.

Follow-up (Table IV)

After a follow-up of 17 to 29 months, the serum WR and VDRL tests were negative in all three cases and the RPCF test also in one. The WR titres fell to zero within 21, 10, and 5 months respectively, and subsequently remained so (Table IV). Thus a steady sero-reversal after therapy provided further evidence of an early syphilitic infection.

Normal or near-normal post-treatment cerebrospinal fluids were obtained in two cases. In the third the cell count was also normal after 8 days but difficulty was experienced with the lumbar puncture and the patient developed 'lumbar puncture spine' which lasted for some months in spite of physiotherapy and a further test was therefore not attempted.

As a result of contact-tracing seven cases of syphilis were found among the consorts of these three men.

Aural recovery

Subjective recovery of discriminative hearing was experienced by all patients who then happily appreciated their music as before.

TABLE IV *Follow-up*

Case no.	1	2	3
Follow-up (mths)	29	21	17
Serology at last visit	WR – VDRL – RPCFT +	WR – VDRL – RPCFT –	WR – VDRL – RPCFT +
Cerebrospinal fluid	Cells 3/c.mm. Protein 39 mg. (per cent.) WR – RPCFT – (5 mths)	Cells 5/c.mm. (8 days) Difficulty experienced; 'lumbar puncture spine'	Cells 1/c.mm. Protein 52 mg. (per cent.) Lange 0000000 WR – RPCFT – (3 and 15 mths)
Contact investigations	17 known contacts (5 positive)	Source S2 5 secondary contacts (2 traced, 1 positive)	5 contacts (1 traced, but was negative)

The objective audiographic pure tone response has so far been the least good in Case 3. The findings apparently worsened within 8 days of starting therapy as a possible Herxheimer effect, and the latest tracing 14 months after therapy, although showing improvement, still indicates some high-tone loss. However, this case has had the shortest follow-up and further improvement may still be possible (Fig. 1).

Case 2 is the most remarkable as showing initially the greatest hearing loss which was further considerably aggravated on the fourth day of treatment as a possible Herxheimer effect of cephaloridine. Subsequent improvement was very slow, but by 21 months was strikingly good (Fig. 2, opposite).

The improvement in Case 1 was perhaps the most satisfactory in so far that it was complete 18 months from treatment (Fig. 3, overleaf).

Summary and conclusions

(1) Syphilitic nerve deafness is commonly considered to be associated with congenital syphilis. Three cases are described in homosexual men who were all concerned with music, and in whom perceptive deafness was considered to have occurred in the early acquired infection. In all three cases the cerebrospinal fluid was abnormal.

(2) All three patients were treated with antibiotics under corticosteroid cover, but in two cases the hearing worsened during the first few days of therapy, in one markedly so, as a probable Herxheimer effect.

(3) After treatment sero-reversal occurred as would be expected in an early infection, and changes towards normality were also observed in the cerebrospinal fluid.

(4) On subjective evidence, recovery of discrimination occurred in all cases and musical appreciation was once more unimpaired. Objective pure tone audiographic measurement showed gradual improvement in one case, almost complete improvement in the second, and a complete return to normality in the third.

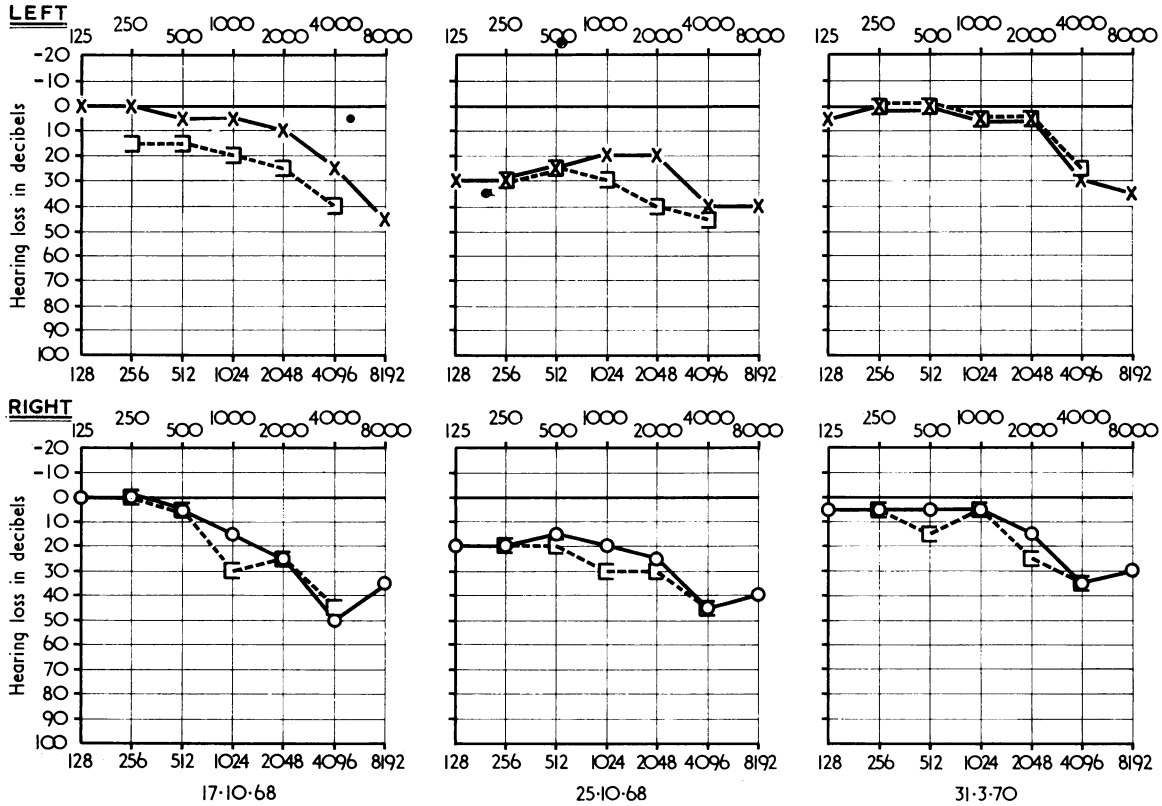

FIG. 1 *Hearing in Case* 1, *right and left ears, on* 17 *and* 25 *October,* 1968, *and* 31 *March,* 1970

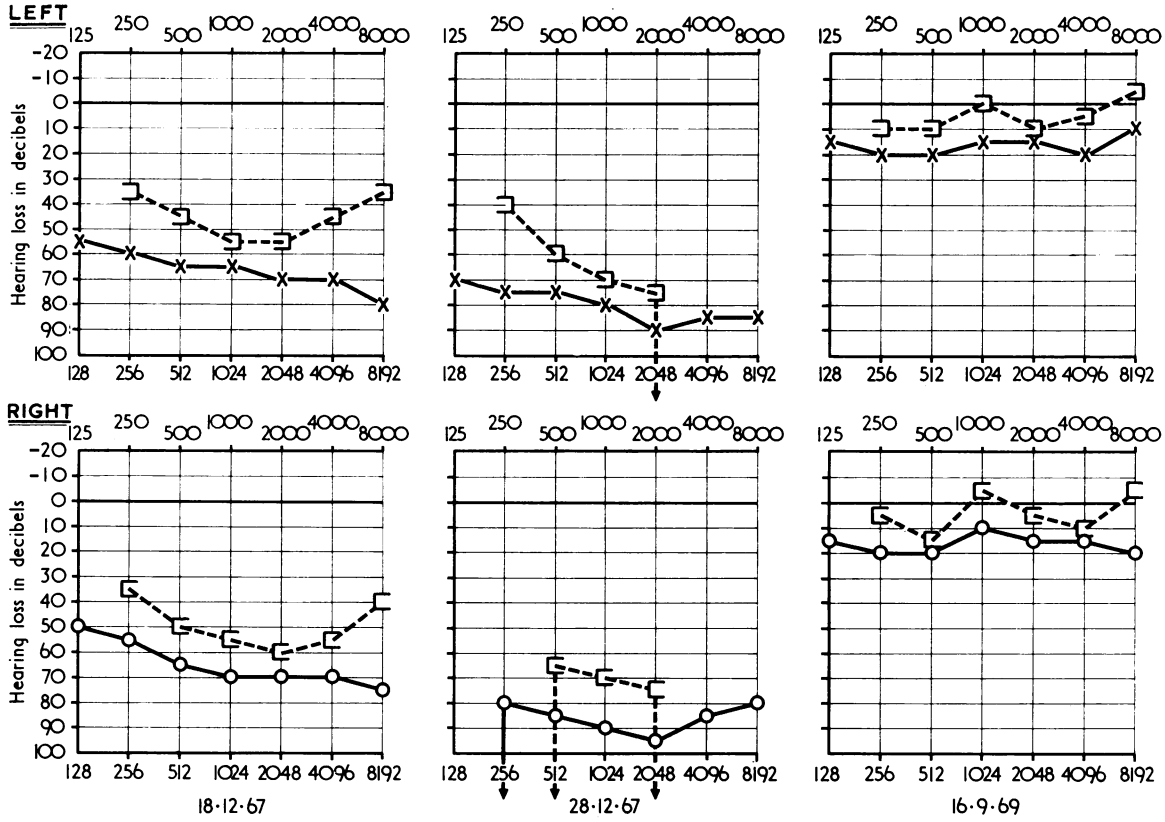

FIG. 2 *Hearing in Case 2, right and left ears, on* 18 *and* 28 *December,* 1967, *and* 16 *September,* 1969

(5) As it was the interest of these patients in music which brought their cases to light, an audiographic study of non-musical secondary syphilitics would be of interest.

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Surdité d'origine nerveuse dans une syphilis récente

SOMMAIRE

(1) On considère généralement que la surdité syphilitique d'origine nerveuse est du domaine de la syphilis congénitale. On rapporte 3 cas d'homosexuels masculins, ayant tous affaire avec la musique, et chez lesquels une surdité de perception fut considérée comme étant survenue au cours d'une syphilis récente acquise. Dans les 3 cas, il y avait des anomalies du L.C.R.

(2) Les 3 malades reçurent un traitement antibiotique sous couverture cortico-stéroïde mais, dans 2 cas, l'acuité auditive se détériora pendant les premiers jours du traitement; ceci fut marqué chez l'un d'eux, probablement par réaction d'Herxheimer.

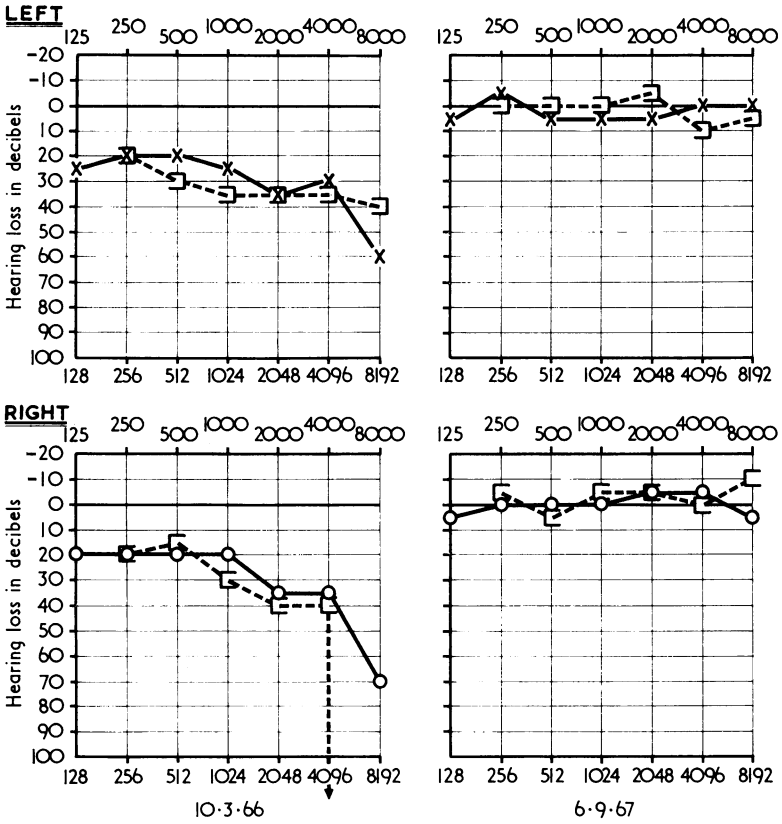

FIG. 3 *Hearing in Case 3, right and left ears, on* 10 *March,* 1966, *and* 6 *September,* 1967

(3) Après le traitement, la sérologie devint négative, comme ceci est habituel dans une infection récente, et le L.C.R. tendit à la normale.

(4) A en juger par les éléments subjectifs, la récupération de la distinction des sons survint dans tous les cas et l'appréciation musicale devint de nouveau correcte. Les mesures audiographiques objectives des sons purs s'améliorèrent graduellement dans 1 cas; l'amélioration fut presque complète pour le second, et le retour à la normale complet pour le troisième. C'est à cause de l'intérêt de cas malades pour la musique que ces cas devinrent apparents; une étude audiographique des non musiciens atteints de syphilis secondaire serait intéressante.